

Pseudoelephantiasis induced by donovanosis

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SUMMARY In a recent survey the incidence of pseudoelephantiasis induced by donovanosis was found to be 5%. The ratio of women to men was 3:1. The incubation period was two to 20 days, and the duration was two to 24 months. The disease was contracted through premarital or extramarital sexual intercourse. The morphological features were characterised in women by globular pedunculated verrucous surfaced swellings affecting the labia majora and the clitoris and accompanying ulceration or its aftermath (depigmentation), or both. Constitutional symptoms were absent. Intracytoplasmic Donovan bodies in the tissue or a smear from the ulcers were diagnostic. Immune markers, such as total T lymphocytes and their subsets (T4 (helper/inducer) and T8 (suppressor/cytotoxic), B lymphocytes, immunoglobulins G, A, and M, and the pivotal complement component C3 were largely inconsequential.

Introduction

Several complications and sequelae of donovanosis are known, pseudoelephantiasis being one of them. The condition was first described by Nair and Pandalai,¹ followed by Rajam and Rangiah.² The reports available on pseudoelephantiasis emphasise that this is an unusual sequel of donovanosis, and its incidence is variable. Most studies in the past were undertaken when treatment was greatly handicapped by the non-availability of specific drugs for the disease. In view of the paucity of recent information on the condition, we decided to undertake a study to establish its current status.

Patients, materials, and methods

We investigated 1090 patients with genital ulcers attending our outpatient clinic during a two year period. When donovanosis was suspected,³ we examined Giemsa stained smears or tissue sections from these ulcers. In addition we performed Gram stained smears of ulcers for *Haemophilus ducreyi*, Giemsa stained smears for giant cells or inclusion bodies of herpes genitalis infection, dark ground microscopy to show *Treponema pallidum*, and serological tests for syphilis, to exclude other causes of ulceration.

Even in morphologically typical ulcers, the ultimate diagnosis of donovanosis was based on demonstrating intracytoplasmic Donovan bodies in large mononuclear cells in Giemsa stained tissue smears or sections.⁴ By these procedures 78 patients were diagnosed as having donovanosis.

We recorded in detail each patient's age, sex, marital and socio-economic status, history of sexual contacts, and incubation period and duration of the disease. Other causes of large genital swellings, such as filaria and lymphogranuloma venereum (LGV) were considered in the differential diagnosis. A history of frequent attacks of fever and an associated increase of swellings indicated filarial infestation, and peripheral blood smears were examined for microfilariae on three consecutive occasions. A history of constitutional symptoms, such as fever, malaise, and arthralgia, invariably preceding or accompanying local manifestations indicated a diagnosis of LGV. Routine haematological and biochemical investigations were undertaken, as was the measurement of numbers of T lymphocytes and their subsets (T4 (helper/inducer) and T8 (suppressor/cytotoxic))⁵ and B lymphocytes and serum concentrations of immunoglobulins⁶ and complement component C3.

Results

Of the 78 patients, three women and one man had pseudoelephantiasis. They were aged between 20 and 30 years, three were married, and one was single. The disease had been contracted through premarital or

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extramarital sexual contact. All patients with pseudoelephantiasis, like most of those with donovanosis, belonged to low socioeconomic strata. One of the women was deaf and dumb, and another had left home during childhood and was a labourer. The duration of this complication varied from two to 24 months, whereas that of the ulcers was from 12 to 30 months. The incubation period of the ulcer varied from two to 20 days. The patients with pseudoelephantiasis had had various local medicines from quacks, and did not report to a major hospital until they had gross swellings.

CLINICAL FEATURES

The women had firm pedunculated globular masses of varying sizes, which affected the labia majora or the clitoris, or both. The overlying skin was oedematous and verrucous with areas of depigmentation (fig 1). Two patients had three swellings, and another had two.



FIG 1 *Pedunculated swellings from the labia majora and the clitoris.*

Ulcers in the intertriginous areas of the swellings were prominent and raised and were formed of soft, granulomatous, friable, and beefy-red tissue (fig 2). The ulcers were tender and bled on touch. The man had gross solid enlargement of distal part of the penis (the overlying skin being oedematous and thickened), phimosis, and an ulcer with typical morphology.



FIG 2 *Elevated soft, friable, granulomatous, beefy-red ulcers in the intertriginous areas.*

In two women routine investigations showed hypochromic microcytic anaemia with simultaneous mild increases in erythrocyte sedimentation rate and concentrations of serum proteins and albumin. Serum concentrations of immunoglobulins were within normal limits. Peripheral blood smears for microfilariae were negative. The tables shows the results of immunological investigations.

Discussion

The incidence of pseudoelephantiasis in the study published here was found to be 5% in patients with donovanosis. This was much lower than in previous studies in which its incidence was reported as 24% by Nair and Pandalai,¹ 15% to 20% by Rajam and Rangiah,² 11% by Rama Rao and Patnaik,⁷ and 13% by Anandam.⁸ The infrequent reporting of pseudoelephantiasis in contemporary published reports may be due to its early diagnosis and specific and adequate treatment of ulcers with tetracyclines,⁹ streptomycin,¹⁰ and recently co-trimoxazole.^{11,12} Our patients with pseudoelephantiasis had chronic disease, with the interval between ulcers and swellings ranging from three to 18 months. Those who fail to get appropriate early and specific treatment may develop this disfiguring complication, and donovanosis should therefore always be considered in the aetiology of elephantiasis of the genitals.

TABLE Results of immunological investigations of four patients with pseudoelephantiasis compared with controls

Variables	Patients		Controls	
	Mean (SD)	Range	Mean (SD)	Range
Differential lymphocyte count (%)	41.8 (6)	34-46	29 (4)	22-36
Total T lymphocytes (%)	68 (3)	65-70	65 (5)	54-73
T4 (helper/inducer) subset (%)	28 (2)	25-30	33 (2)	27-38
T8 (suppressor/cytotoxic) subset (%)	19 (5)	12-25	20 (4)	19-26
B lymphocytes (%)	25 (3)	20-28	18 (3)	14-24
Immunoglobulin G (g/l)*	20 (10)	19.8-20.0	12.5 (0.2)	9.50-16
Immunoglobulin A (g/l)	2.4 (0.5)	1.8-3.0	2.2 (0.6)	1.35-3.5
Immunoglobulin M (g/l)	2.2 (0.6)	1.5-2.8	1.7 (0.4)	1.1-2.8
Complement component C3 (g/dl)	1.2 (0.3)	1.7 (0.4)	1.2-2.8	
	0.8-1.5			

* Increased concentrations of IgG indicates chronicity of the disease.

The precise pathogenesis of pseudoelephantiasis caused by donovanosis is still obscure. The mechanical pressure of scar tissue over the local lymphatics coupled with a continuing chronic inflammatory response may be responsible for the condition, which may be influenced by the duration of the donovanosis. Evaluation of the immune responses was not helpful.

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